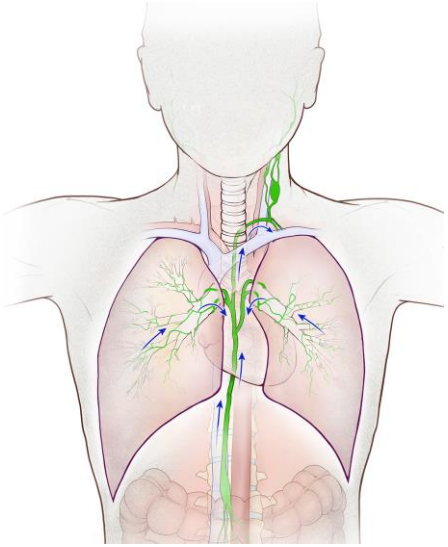


Perelman School of Medicine
University of Pennsylvania

Interventional Management of Lymphatic Morbidity in Patients With CHD

Maxim Itkin MD, FSIR

Professor of Radiology and Pediatrics

Hospital of University of Pennsylvania

DISCLOSURE STATEMENT OF FINANCIAL INTEREST

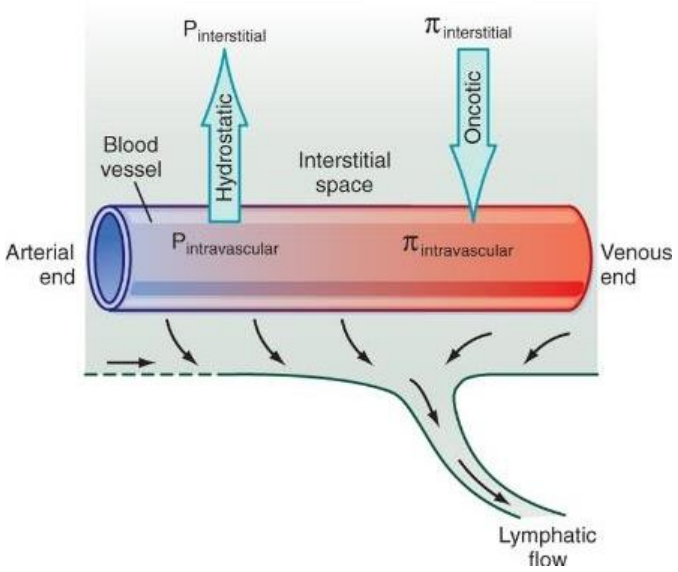
Within the past 12 months, I or my spouse/partner have had a financial interest/arrangement or affiliation with the organization(s) listed below

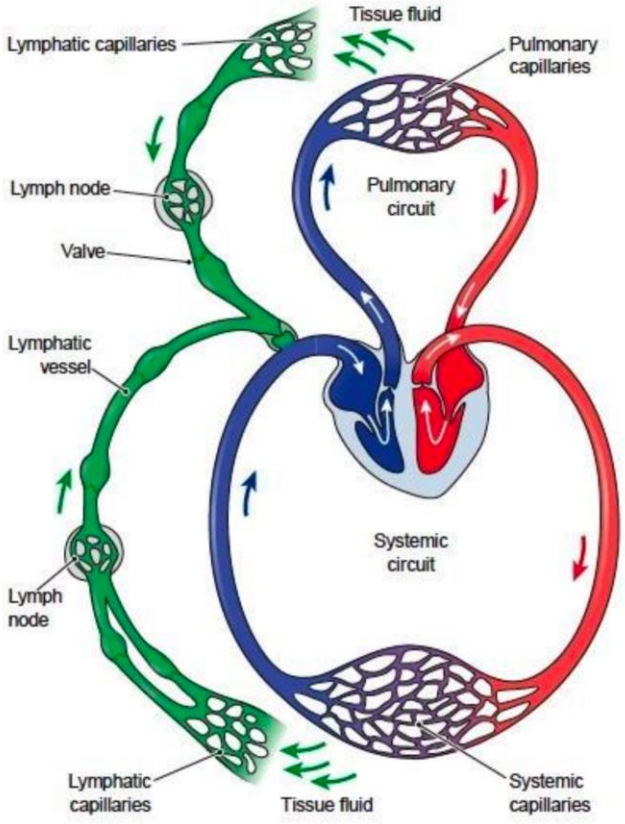
AFFILIATION/FINANCIAL RELATIONSHIP	COMPANY
• Grant/Research Support	• Guerbert Group
• Consulting Fees/Honoraria	• Cook inc
• Ownership/Founder	• Controlrad

Lymphatic System Function

Removal of the transudate (proteins/water)

from the tissues back into systemic circulation

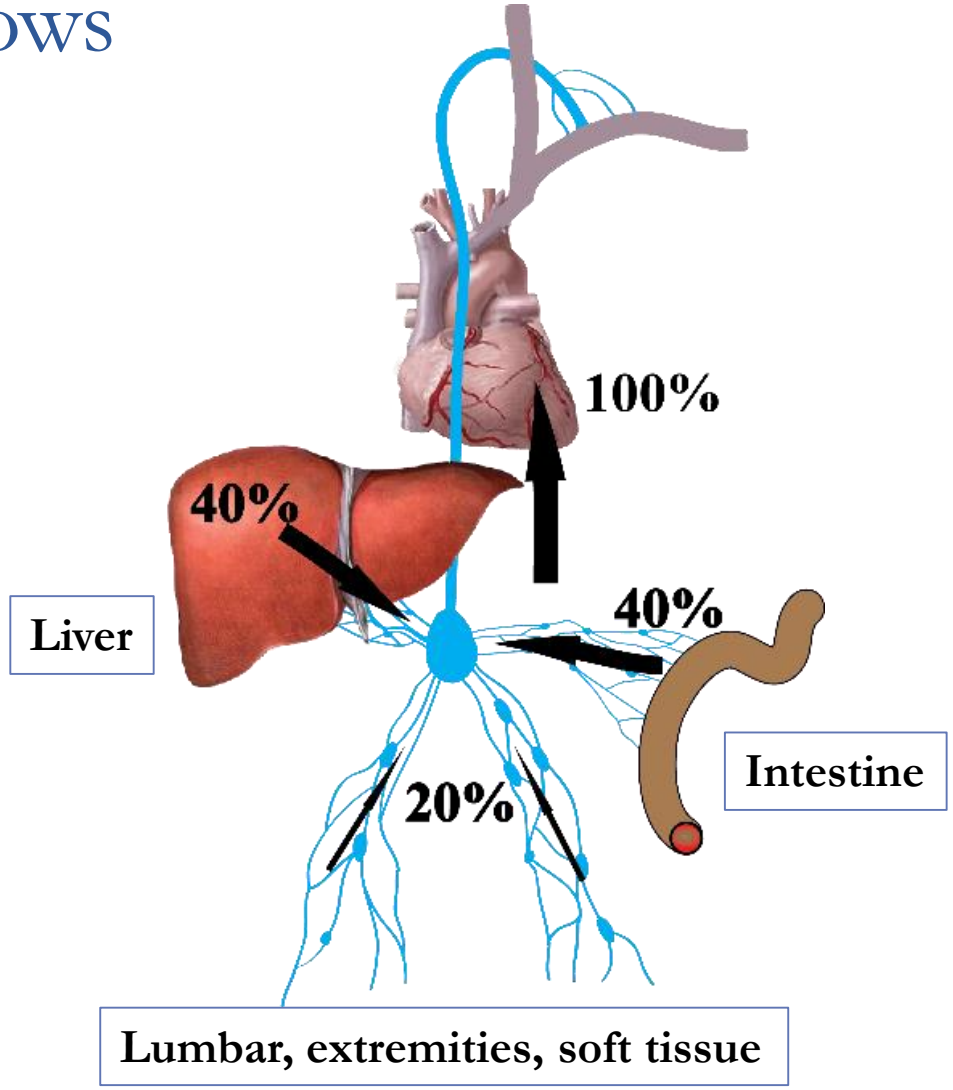




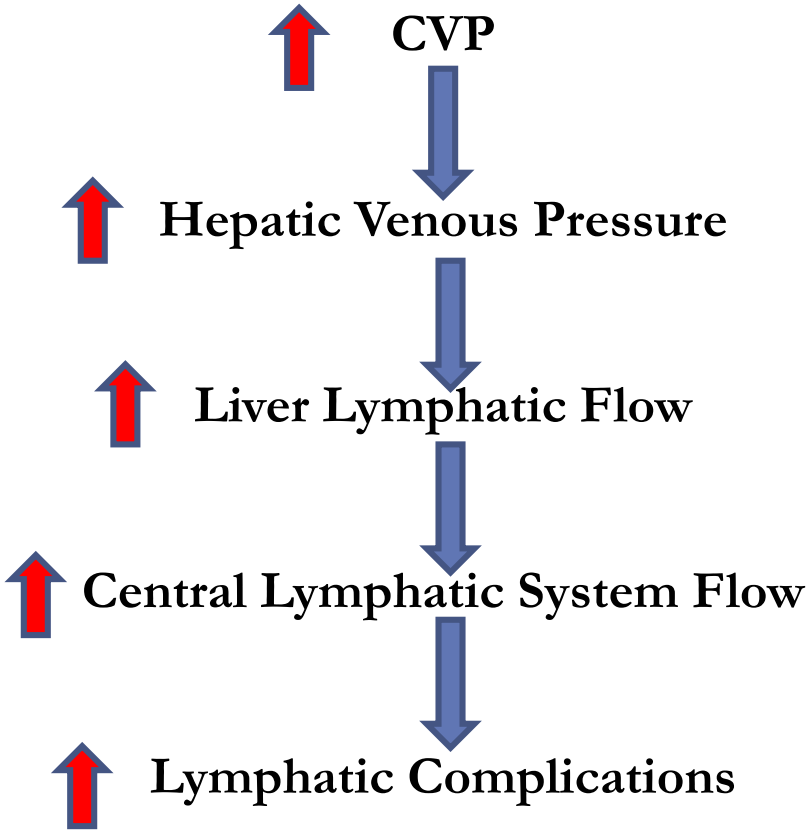
Lymphatic System-Flows

Lymphatic Systems

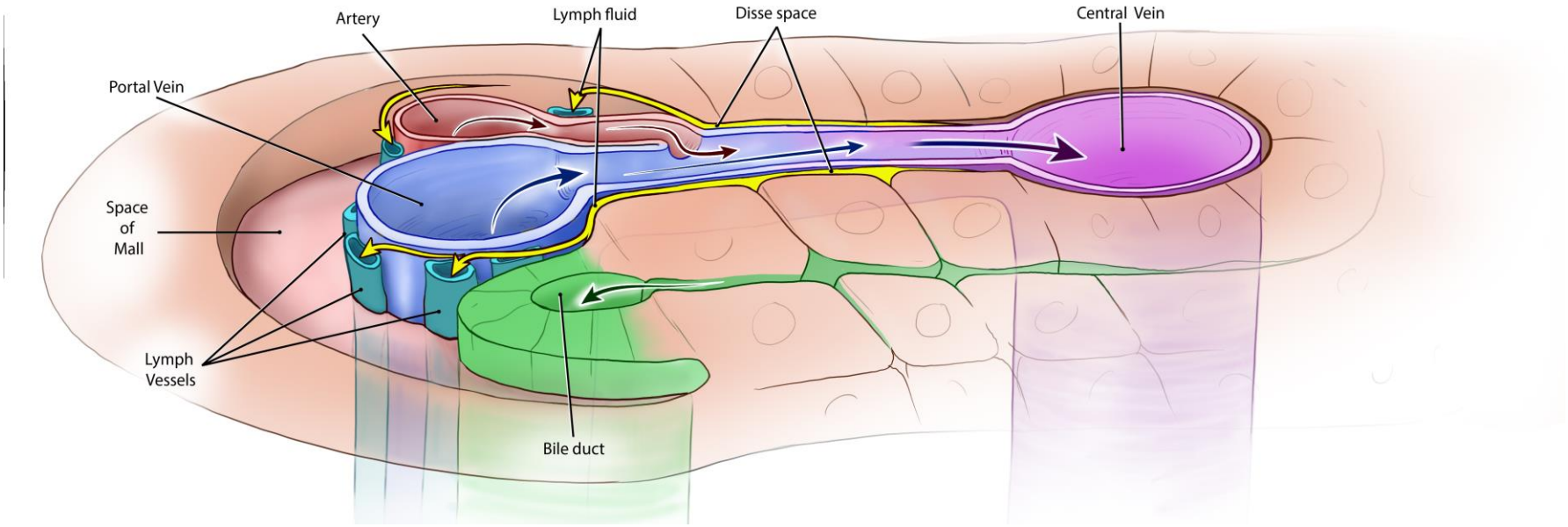
- Liver
- Intestine
- Soft tissue



Lymphatic Consequences of Congenital Heart Surgery



Liver Lymph Production



Pressure in the sinusoid is low
Need to maintain positive influx of ultrafiltrate
Hepatic sinusoids are highly permeable to albumin

Ernest Starling-1887

It seems probable that the obstruction to flow of lymph from the thoracic duct into the blood as well as the distention of the duct from the largely increased lymph flow from the liver which is present in uncompensated heart disease may contribute to the production of edema in the rest of the body (and) to the production of hydrothora ate

ErnestH. Starling, The Arris and Bale lectures on some points]n the pathology of heart disease., In The Lancet, Volume 149, Issue 3835, 1897, Pages 569-572

Ernest Starling-1897

It seems probable that the obstruction to flow of lymph from the thoracic duct into the blood as well as the distention of the duct from the largely increased lymph flow from the liver which is present in uncompensated heart disease may contribute to the production of edema in the rest of the body (and) to the production of hydrothora ate

Ernest H. Starling, The Arris and Bale lectures on some points]n the pathology of heart disease., In The Lancet, Volume 149, Issue 3835, 1897, Pages 569-572

Size of the Thoracic Duct in Heart Failure

Congestive Heart Failure on MRL

Dori, Y., Keller, M. S., Fogel, M. A., Rome, J. J., Whitehead, K. K., Harris, M. A., & Itkin, M. (2014). MRI of Lymphatic Abnormalities After Functional Single-Ventricle Palliation Surgery. *AJR Am J Roentgenol*, *203*(2), 426–431..

Lymphatic Complications of Congenital Heart Surgery

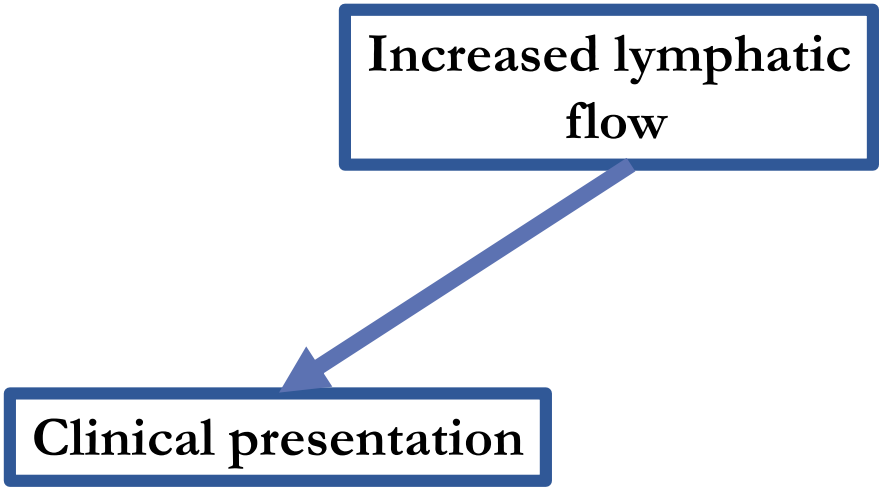
- **Abnormal Pulmonary Lymphatic Flow**
  - Postsurgical Chylothorax
  - Plastic Bronchitis
- **Abnormal Hepatoduodenal Lymphatic Flow**
  - Protein Losing Enteropathy

Lymphatic Complications of Congenital Heart Surgery

There is lack of correlation between the severity of heart failure and incidence of lymphatic complications

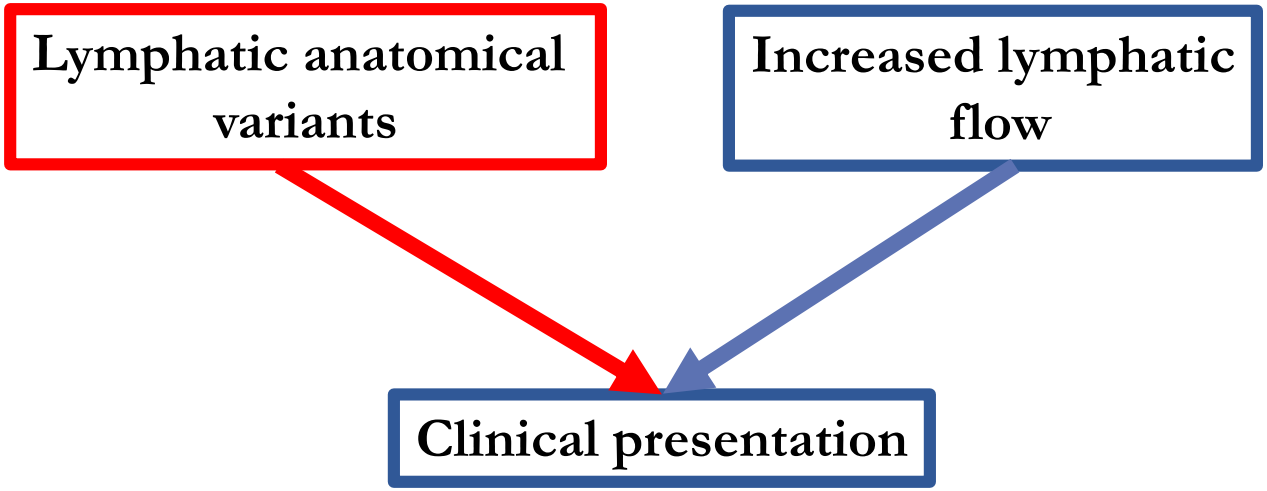
Lymphatic Complications of Congenital Heart Surgery

There is lack of correlation between the severity of heart failure and incidence of lymphatic complications



Lymphatic Complications of Congenital Heart Surgery

There is lack of correlation between the severity of heart failure and incidence of lymphatic complications

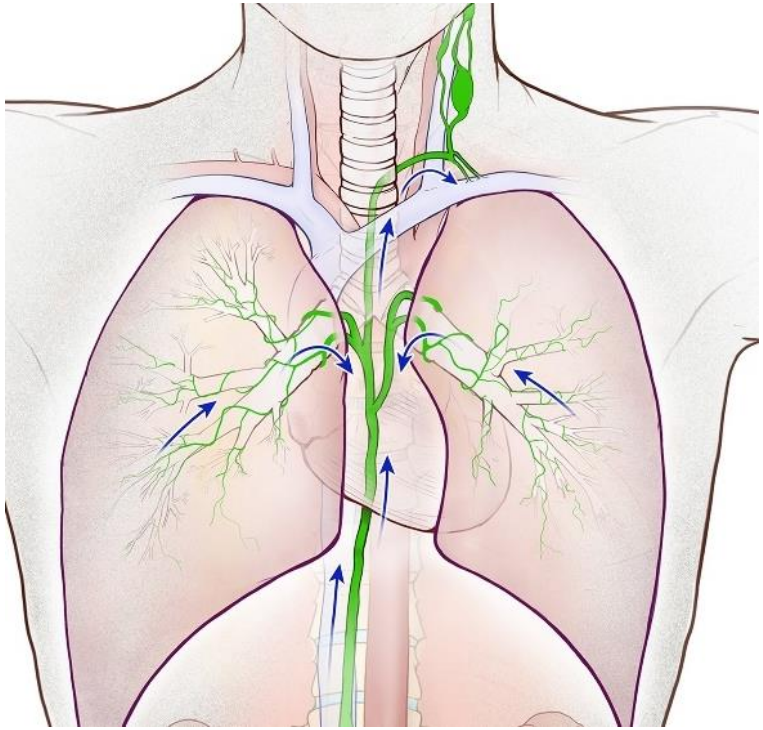


Abnormal Pulmonary Lymphatic Perfusion

Congenital Lymphatic Variant

Plastic Bronchitis

Post Cardiac Surgery Chylothorax



Abnormal Pulmonary Lymphatic Perfusion

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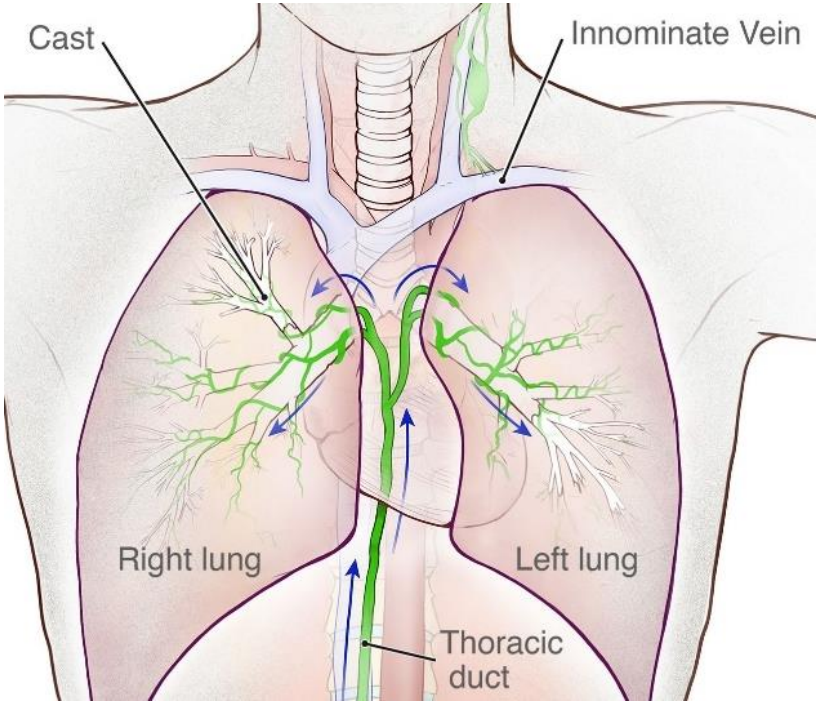
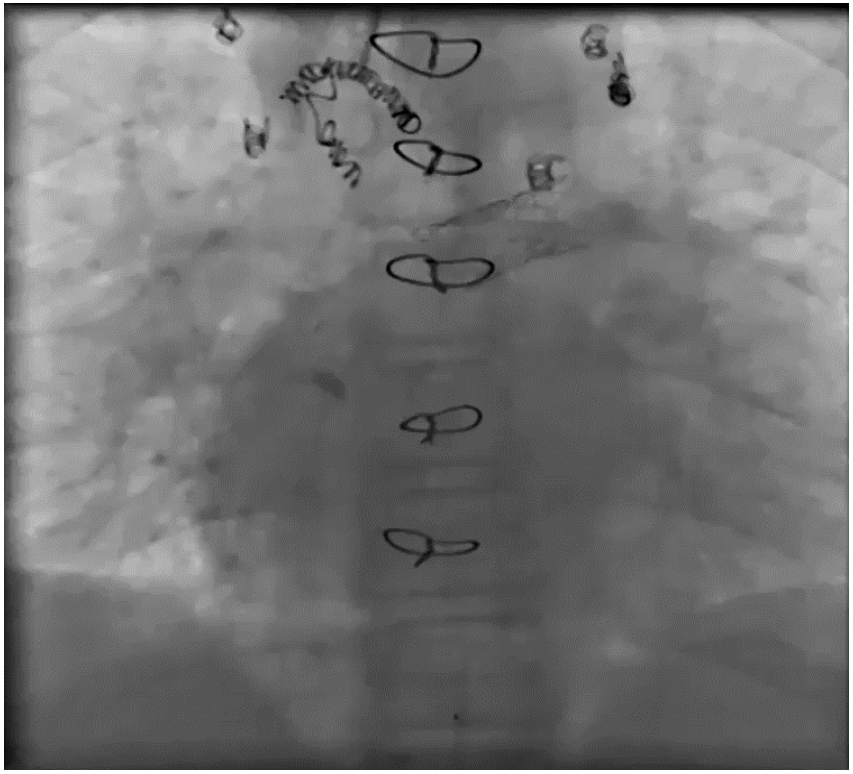


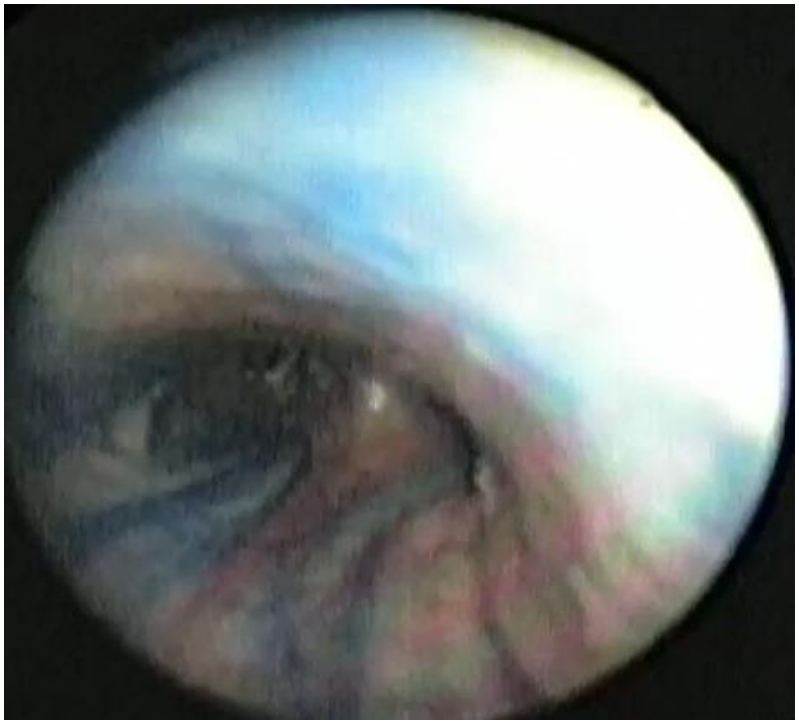
Plastic Bronchitis

- **Formation of large gelatinous or rigid branching airway casts**

Lymphangiography



TD Injection with Methylene Blue



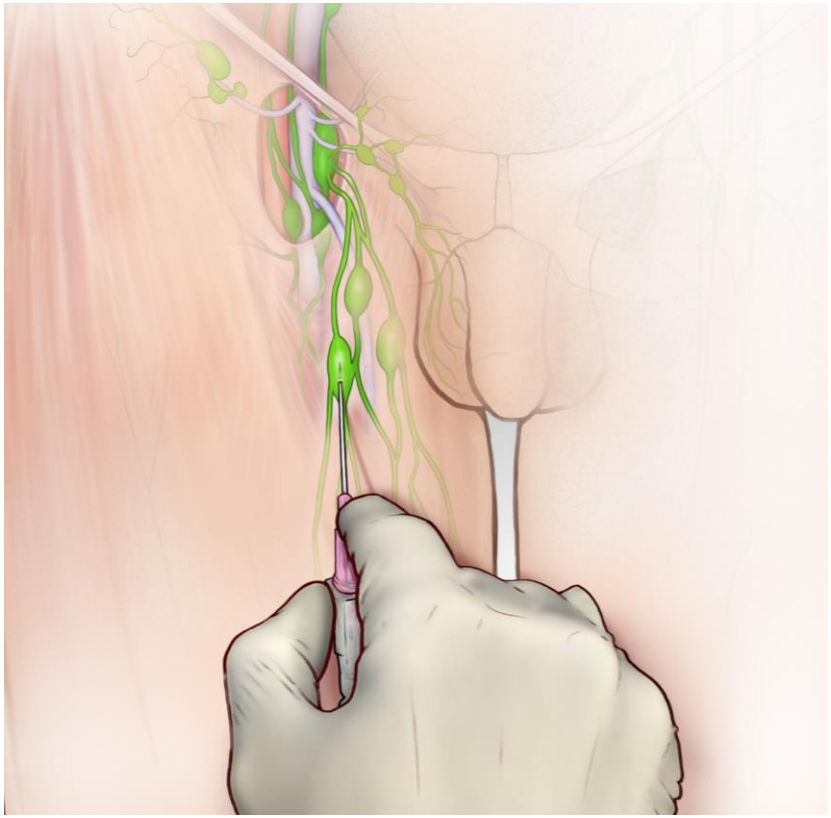
Treatment - TD Embolization

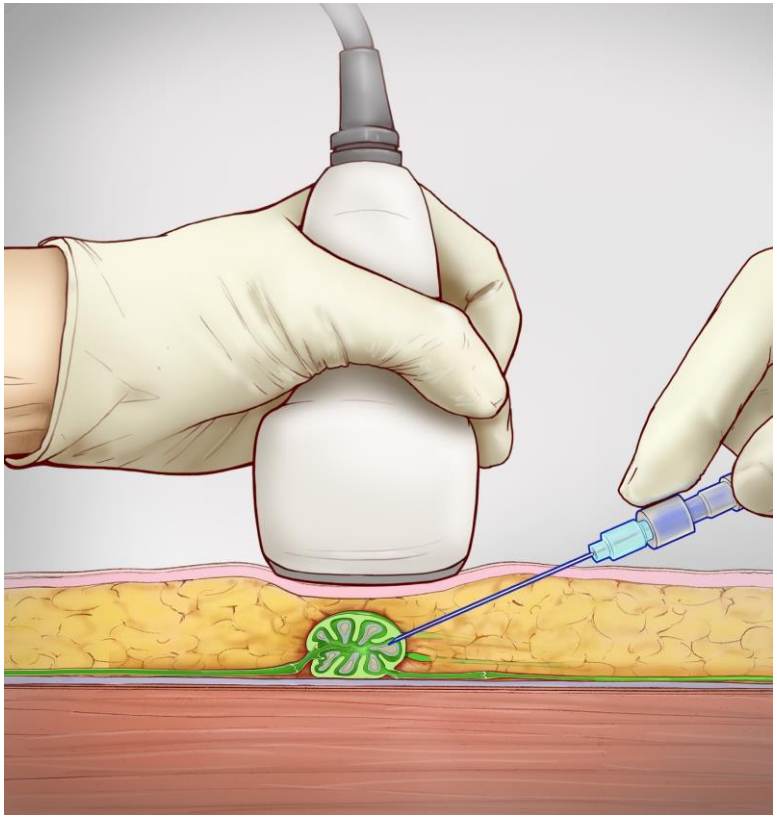
Intranodal Lymphangiography

TD catheterization

TD embolization

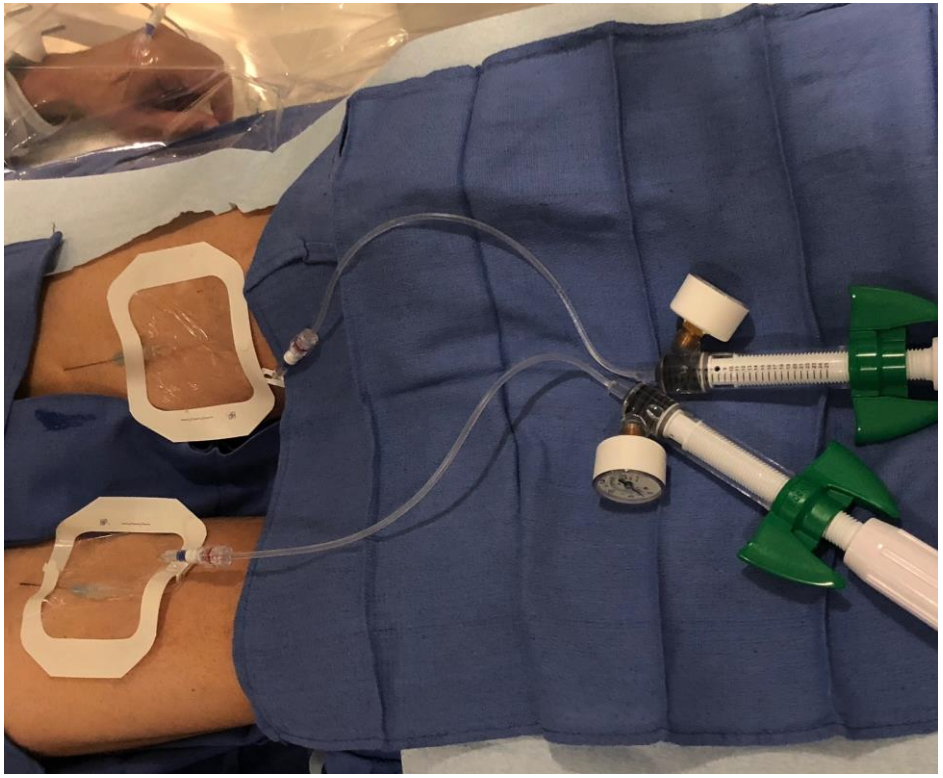
Intranodal Lymphangiogram





Nadolski GJ, Itkin M. Feasibility of ultrasound-guided intranodal lymphangiogram for thoracic duct embolization. J Vasc Interv Radiol. 2012

Intranodal Lymphangiogram



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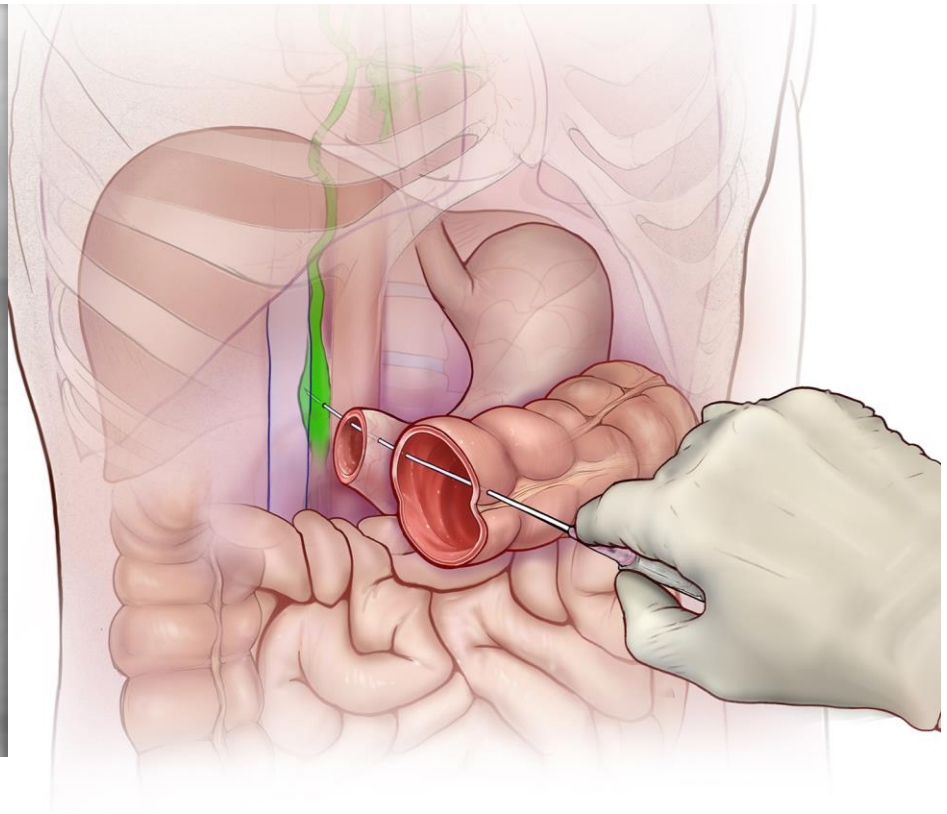
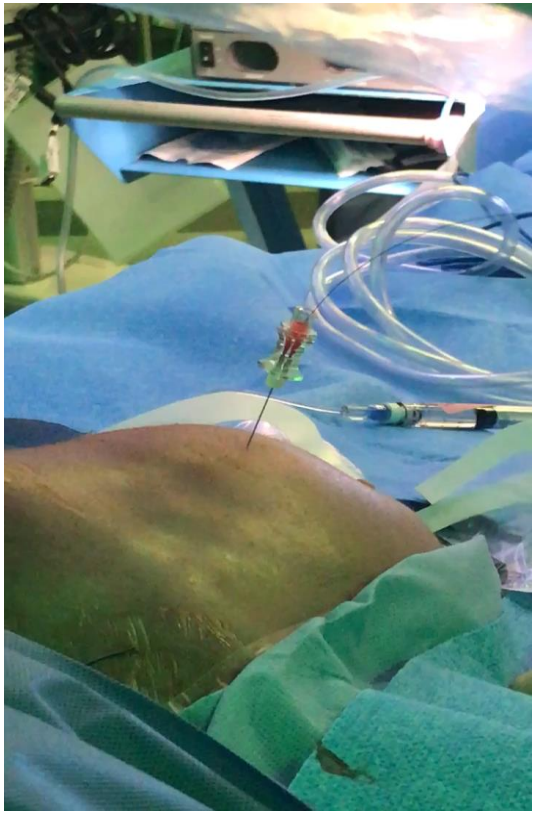
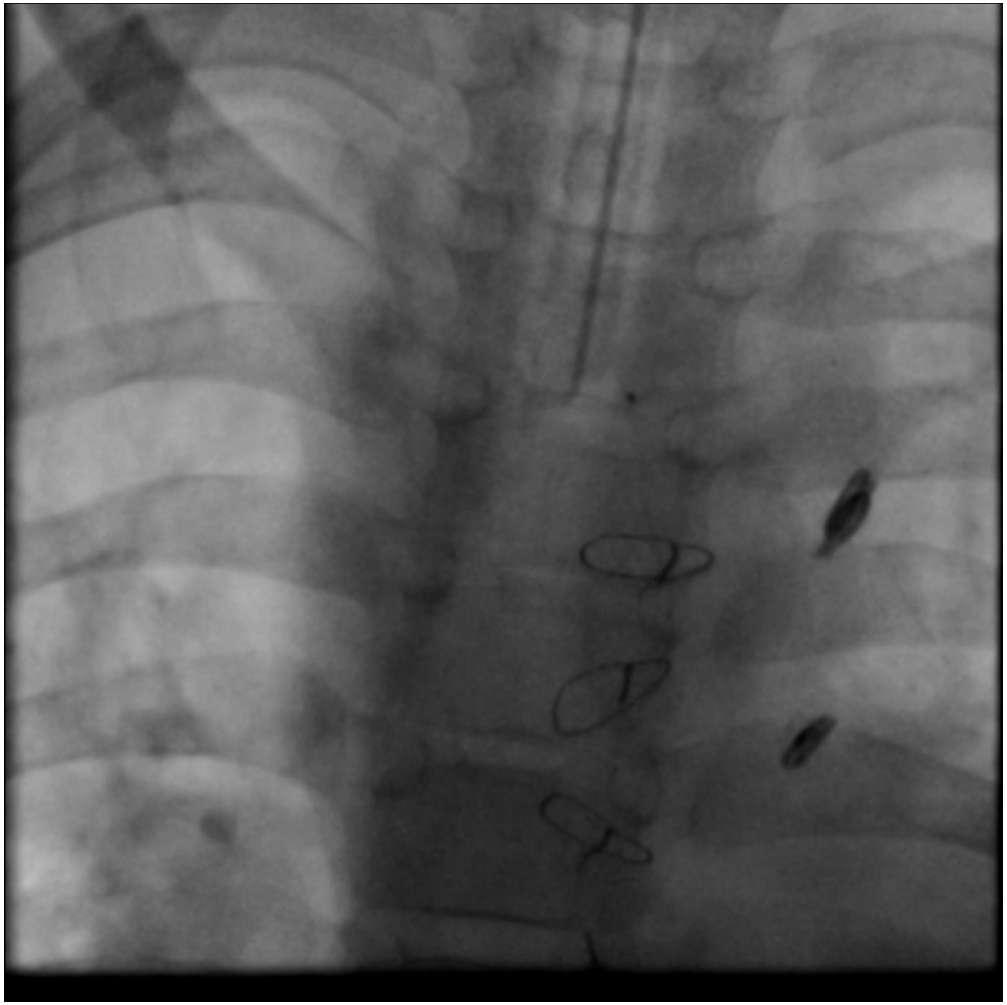
Thoracic Duct Access



Thoracic Duct Access



TD Injection



Embolization

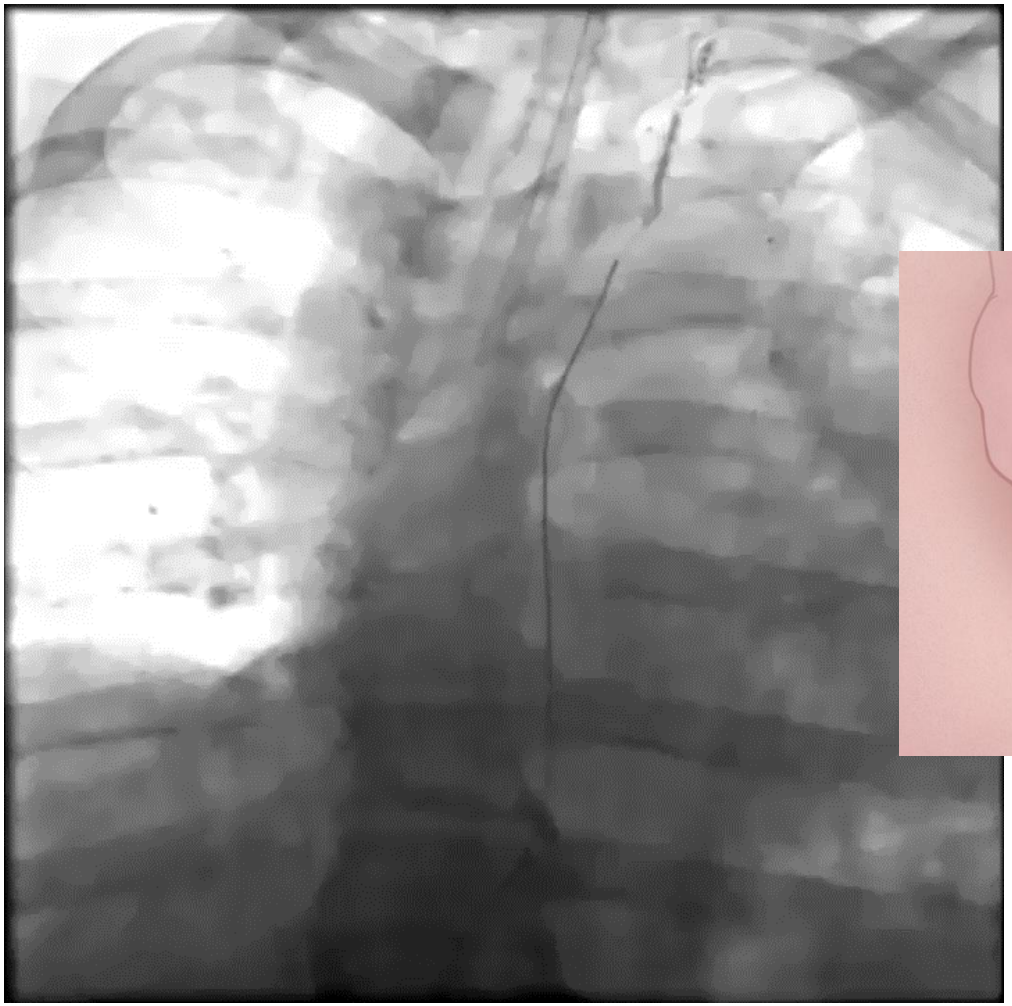
Microcoils

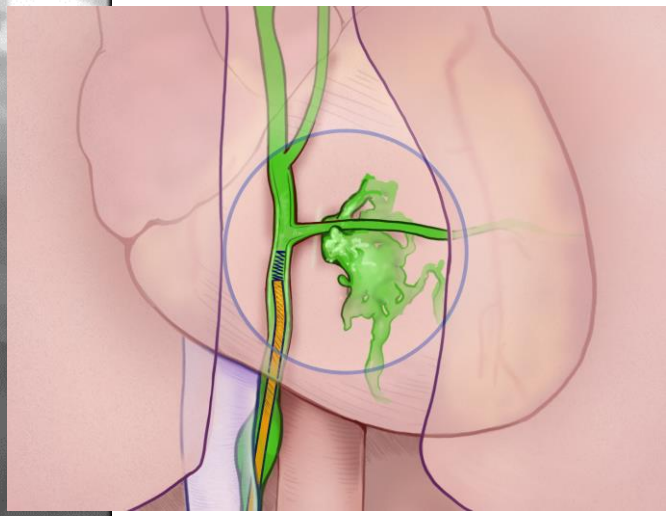
- Nester 0.018

Liquid embolic agent

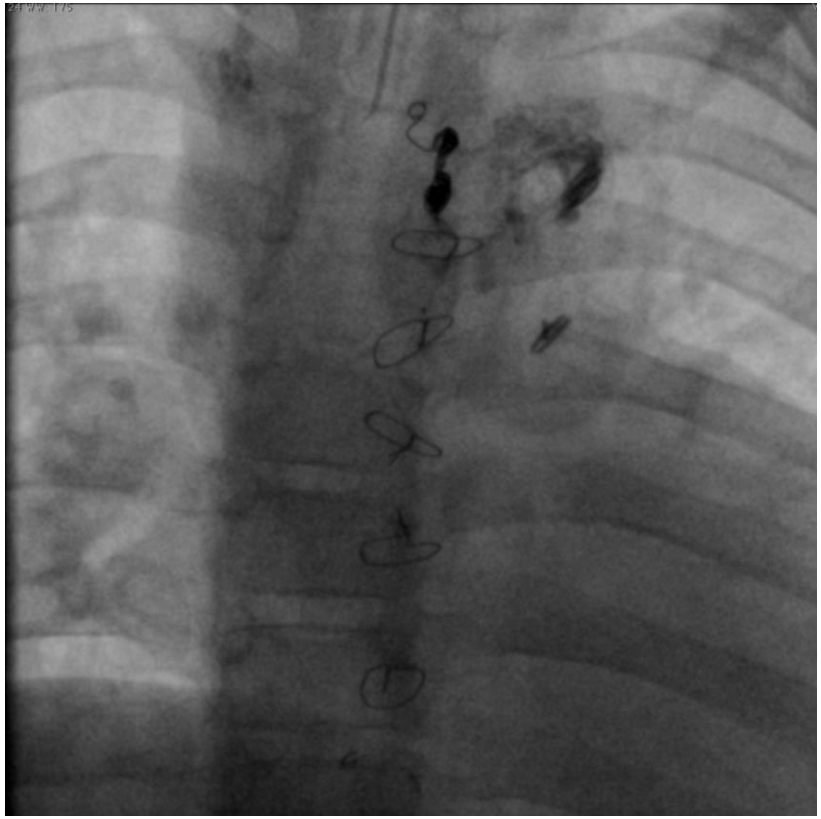
- Glue -n-Butyl Cyanoacrylate (n-BCA)

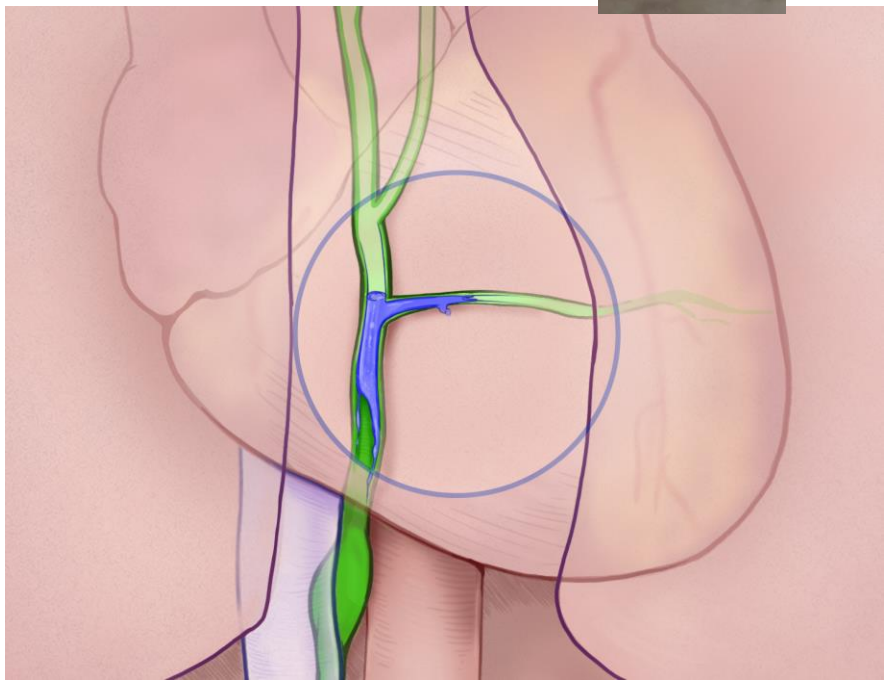
Coil Deployment





Glue injection





PB Lymphatic Embolization-Outcome

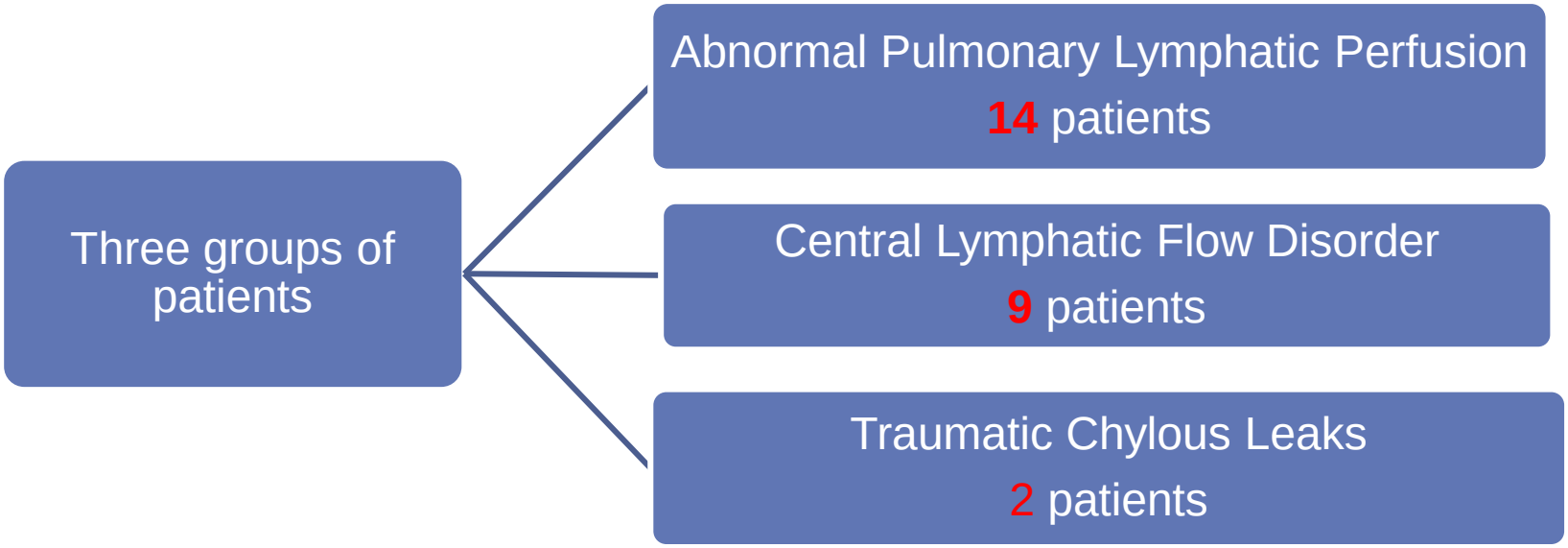
- **18 Patients with "cardiac" PB**
- **16 demonstrated pulmonary lymphatic perfusion**
- **15/16(94%) –significant improvement of their symptoms**
- **Median follow-up 315 days**
- **One major complication-TIA**
  - Lymphatic to pulmonary vein

Y Dori, M Keller, JJ Rome, MJ Gillespie, AC Glatz, K Dodds, DJ Goldberg, S Goldfarb, J Rychik, M Itkin
Percutaneous Lymphatic Embolization of Abnormal Pulmonary Lymphatic Flow as Treatment of Plastic Bronchitis in Patients With Congenital Heart Disease. Circulation 2016;133:1160–1170.

Post Pediatric Cardiac Surgery Chylothorax

Retrospective review of 25 patients

MR lymphangiography and Intranodal Lymphangiography

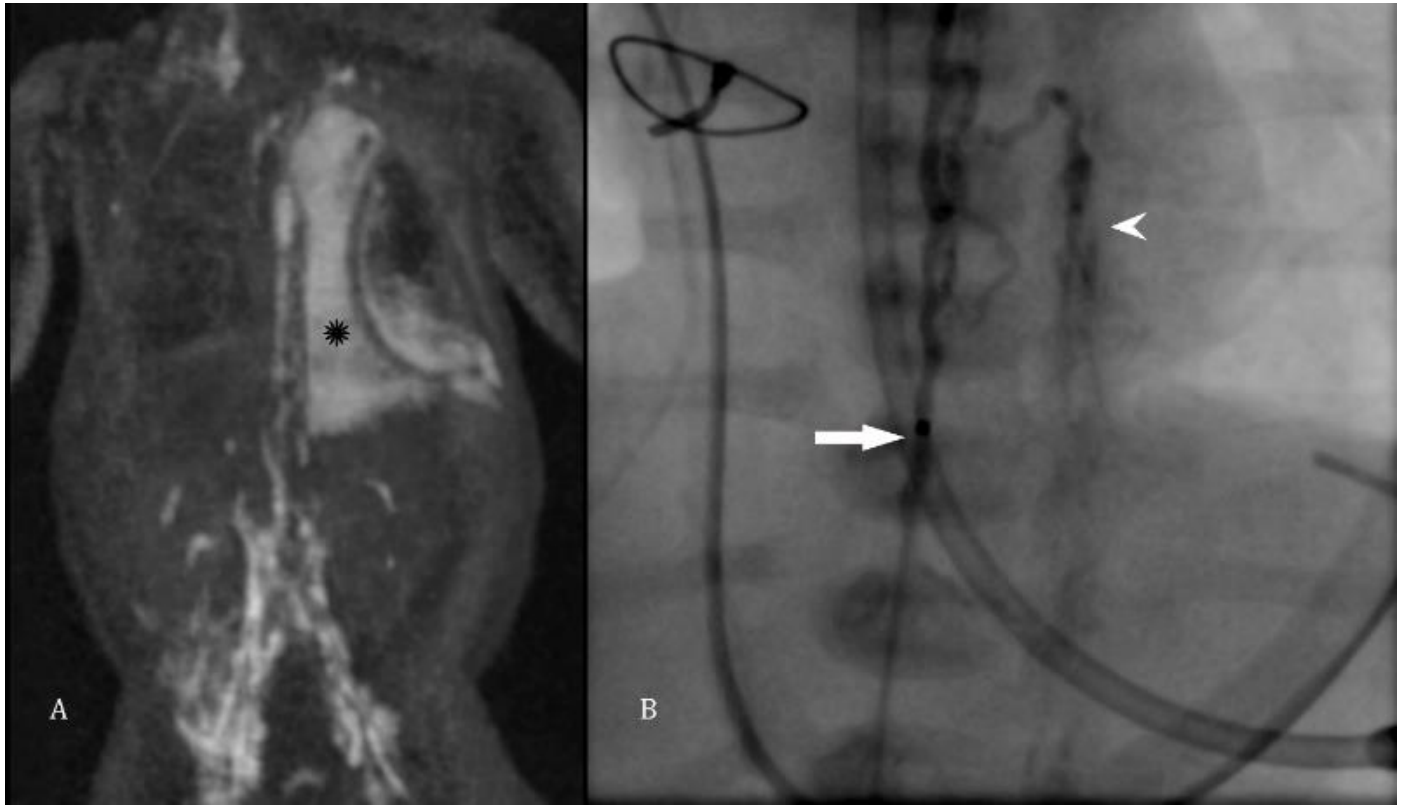


Savla, J. Itkin M et al (2017). Post-Operative Chylothorax in Patients With Congenital Heart Disease. *Journal of the American College of Cardiology*, *69*(19), 2410–2422.

Pulmonary Lymphatic Perfusion Syndrome

Savla, J. Itkin M et al (2017). Post-Operative Chylothorax in Patients With Congenital Heart Disease. *Journal of the American College of Cardiology*, *69*(19), 2410–2422.

Traumatic Chylous Leaks



Savla, J. et al (2017). Post-Operative Chylothorax in Patients With Congenital Heart Disease. *Journal of the American College of Cardiology*, *69*(19), 2410–2422.

Central Lymphatic Flow Disorder

Absent/Diminutive TD

Combination of ascites and pleural effusion

Tissue edema

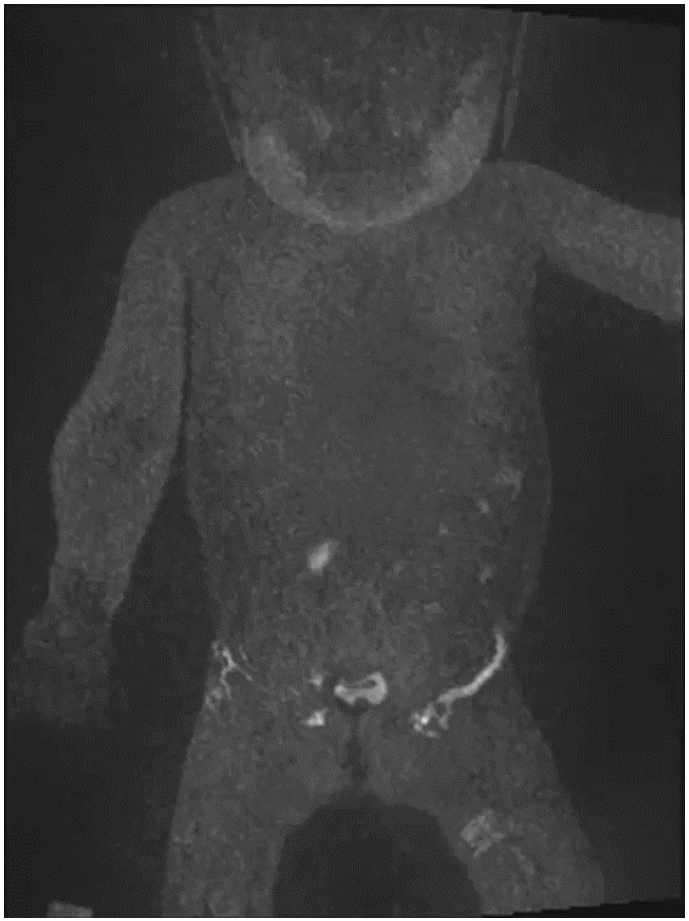
Dermal collaterals

Under 1 year of age

4/9 patient had TD ligation

Savla, J. Itkin M et al (2017). Post-Operative Chylothorax in Patients With Congenital Heart Disease. *Journal of the American College of Cardiology*, *69*(19), 2410–2422.

Central Lymphatic Flow Disorder



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Results

PLPS and Trauma - 16 patients

- TD embolization-all patients
- All 16 patients had resolution of chylothorax after embolization

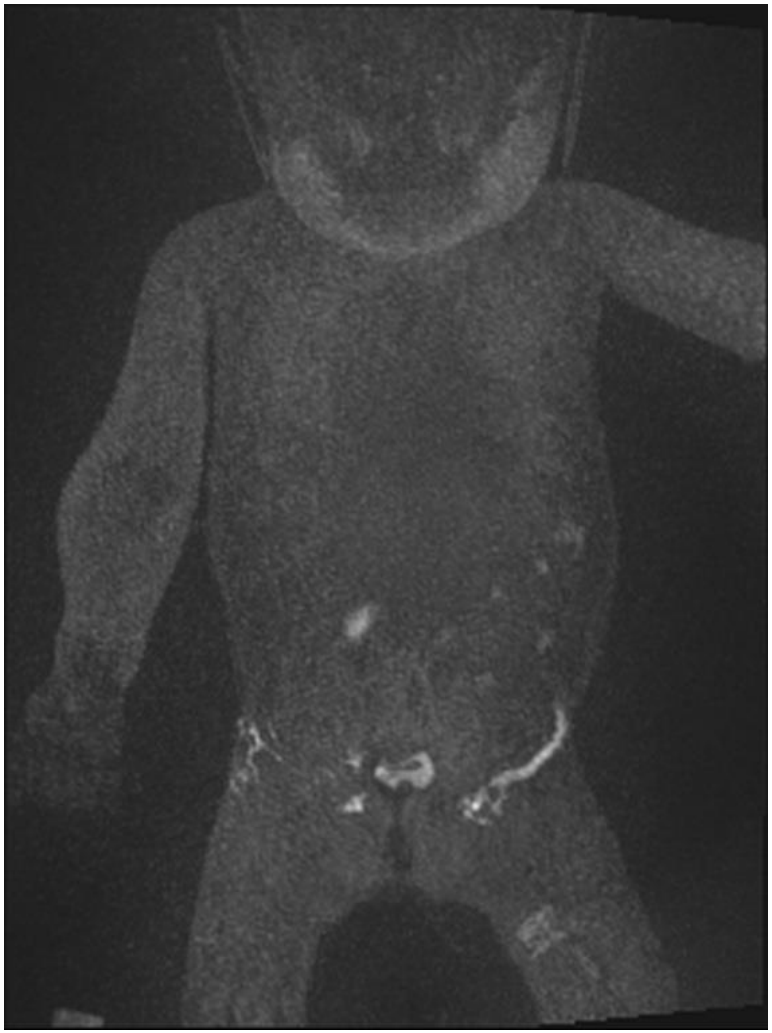
Central Lymphatic Flow Disorder - 9 patients

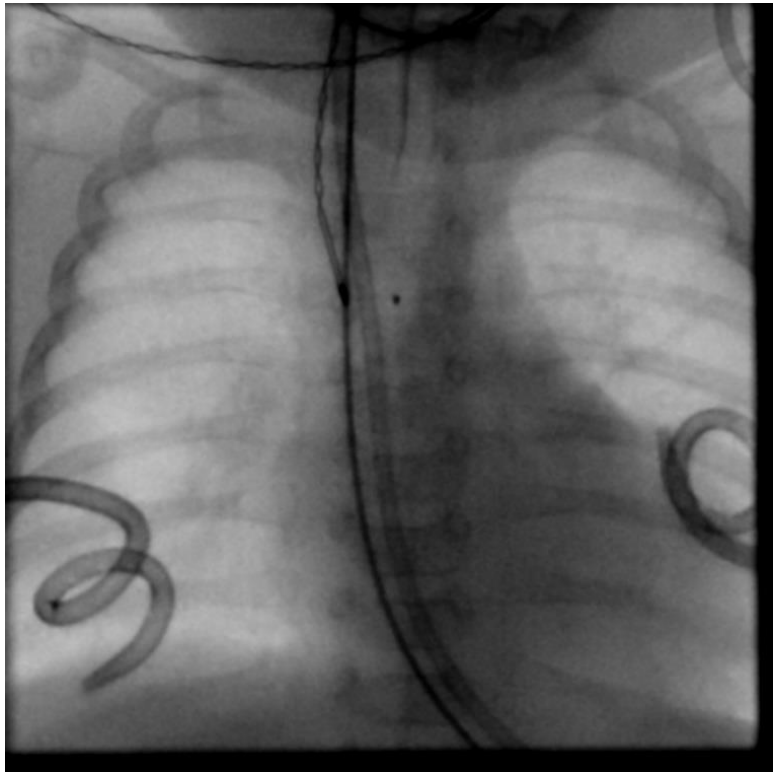
- TD embolization- 6 patients
  - Clinically unsuccessfully
- One patient underwent lympho-venous anastomosis
  - Resolution of chylothorax
- 7 patients deceased

Liver

Savla, J. Itkin M et al (2017). Post-Operative Chylothorax in Patients With Congenital Heart Disease. *Journal of the American College of Cardiology*, *69*(19), 2410–2422.

Central Lymphatic Flow Disorder





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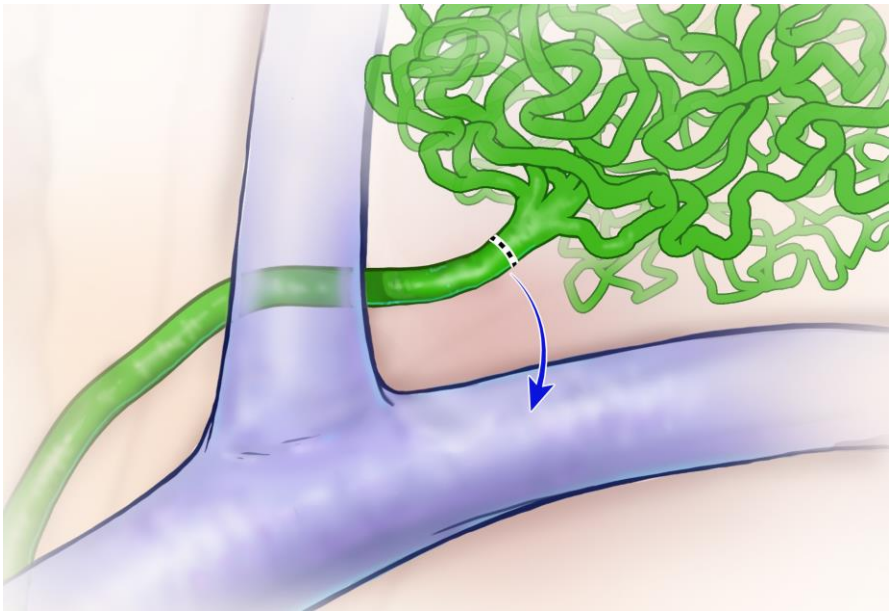
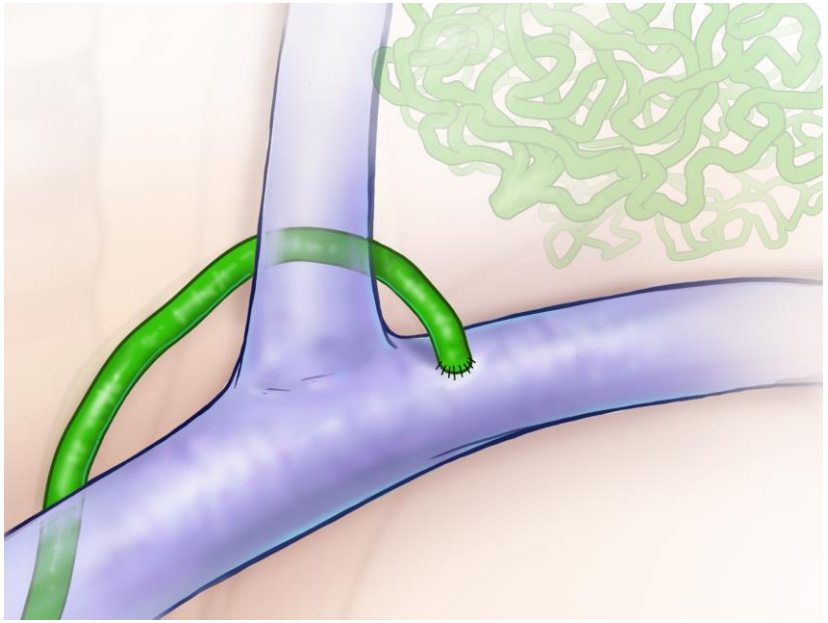
TD-Venous Anastomosis





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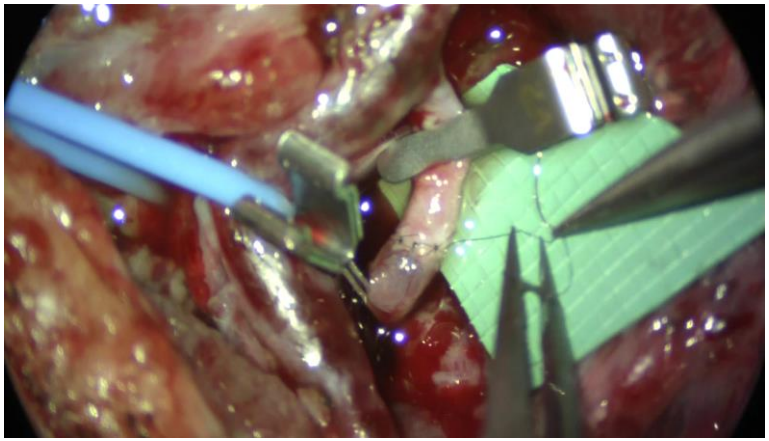
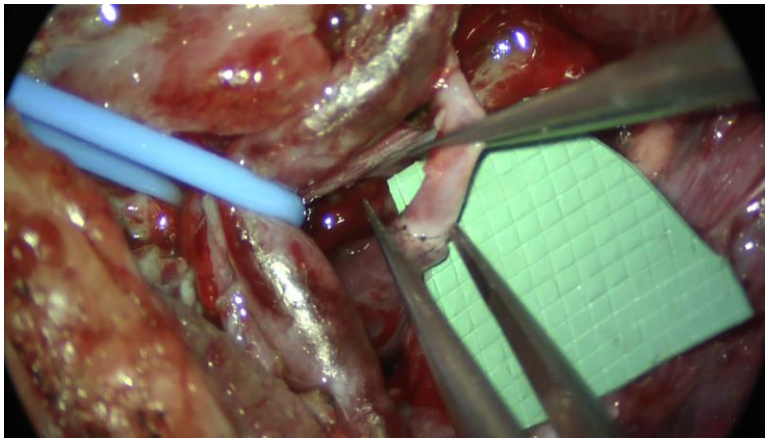
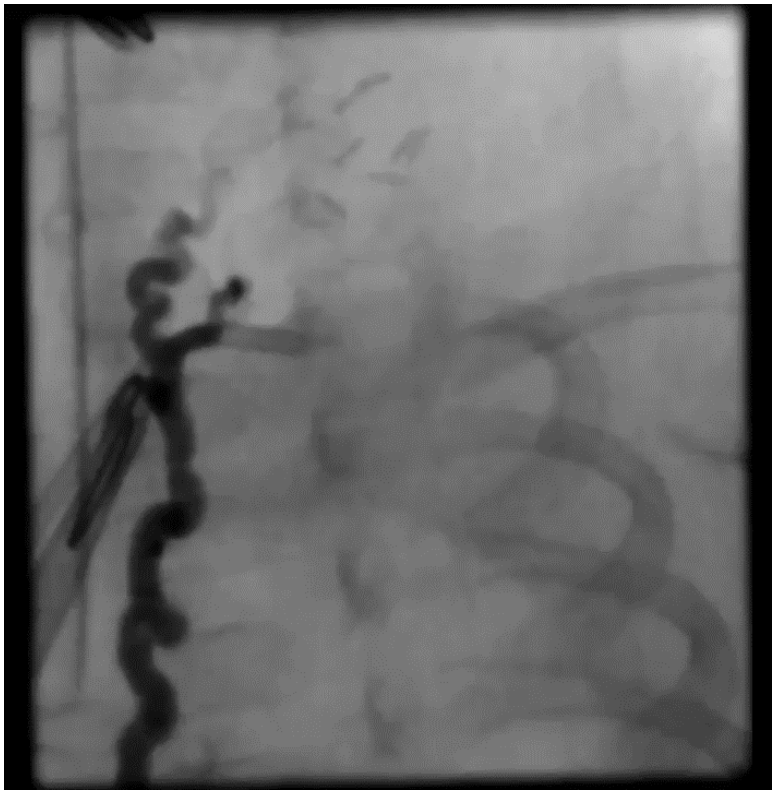
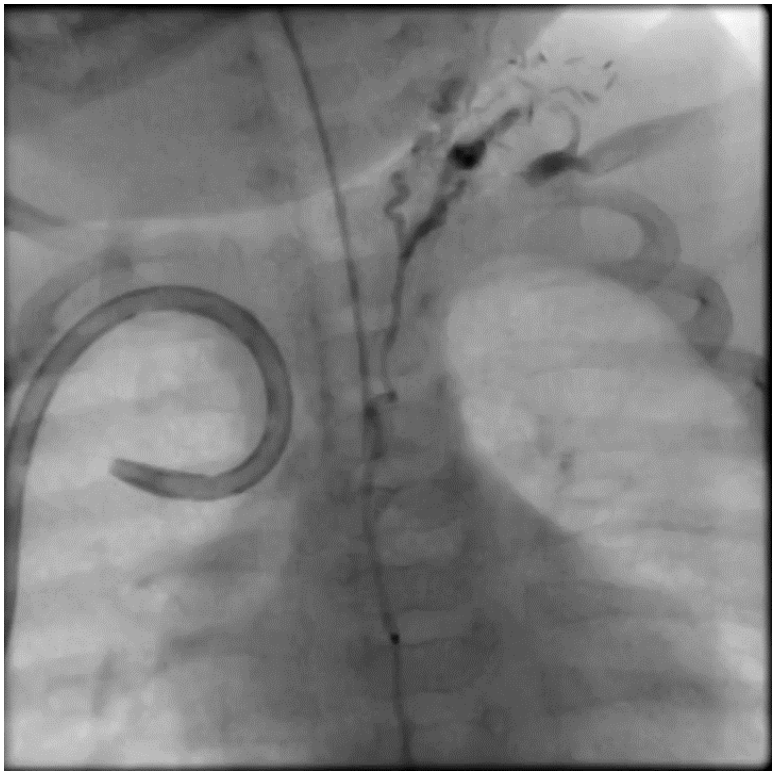


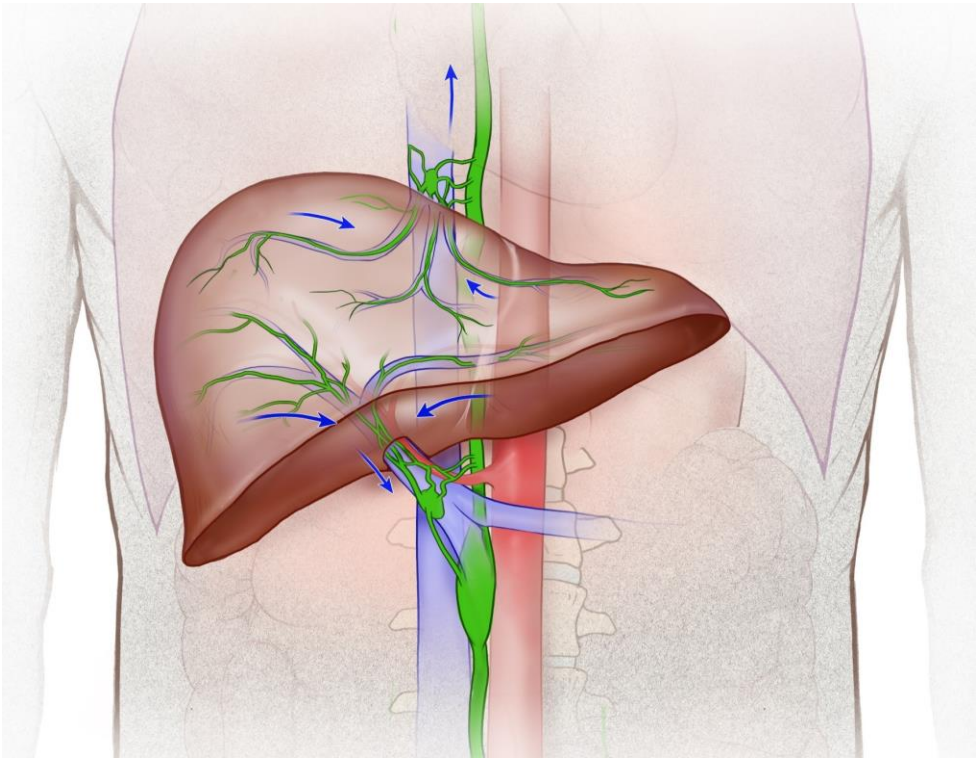


TD-Venous Anastomosis



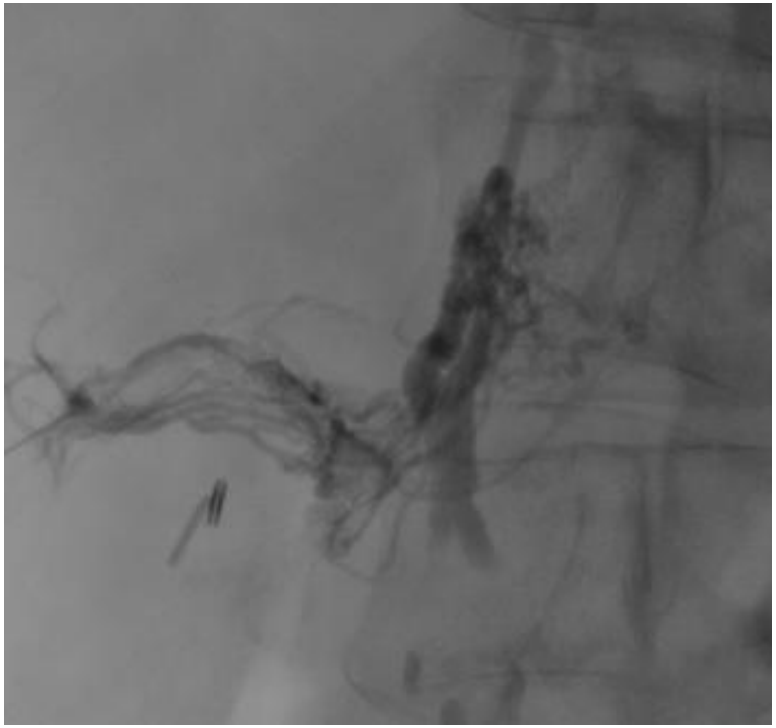


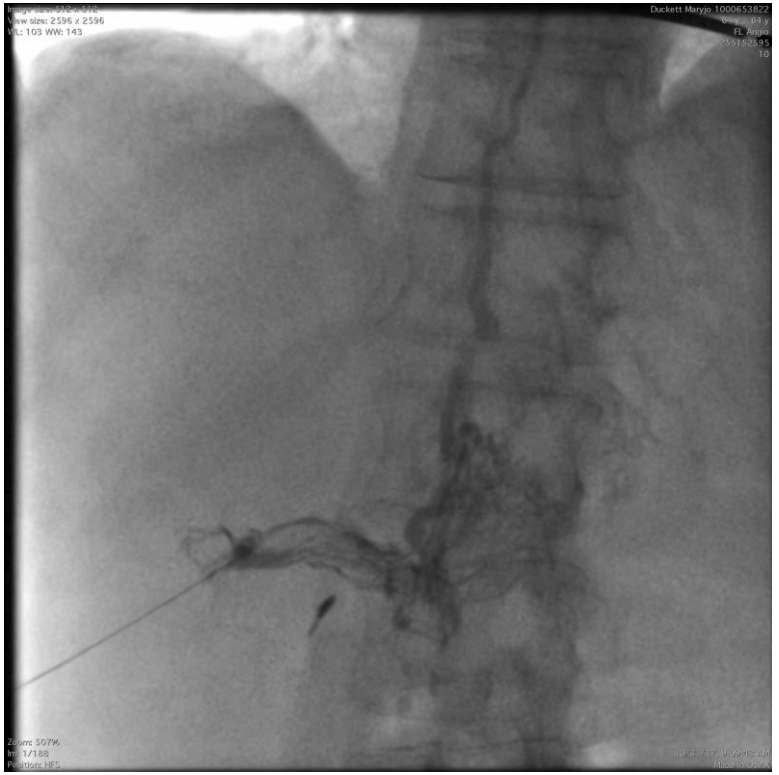
Liver Lymphatic System-Anatomy



- **Liver lymph 40% of the flow in TD**
- **High concentration of the albumin**
- **Flow increases significantly in heart failure**

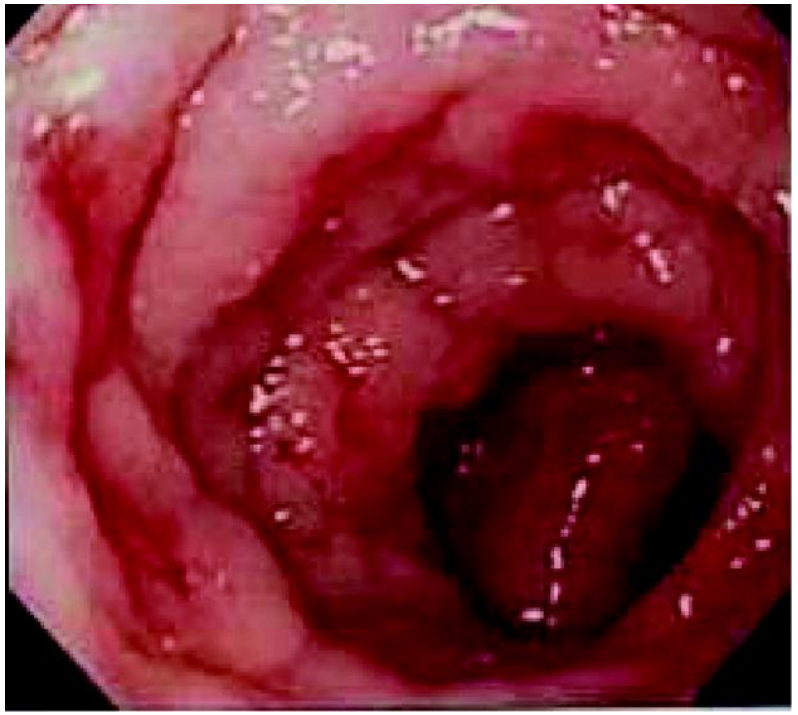
Communication of the Liver Lymphatics with TD





Protein Losing Enteropathy

- **Severe loss of serum proteins into the intestine**



PLE Pathophysiology Concept

Physiology :

- Liver generates albumin and delivers it into the blood stream through lymphatic system
- Liver lymph has a high concentration of proteins
- The lymphatic flow in the liver increases significantly in patients with CHF

Hypothesis:

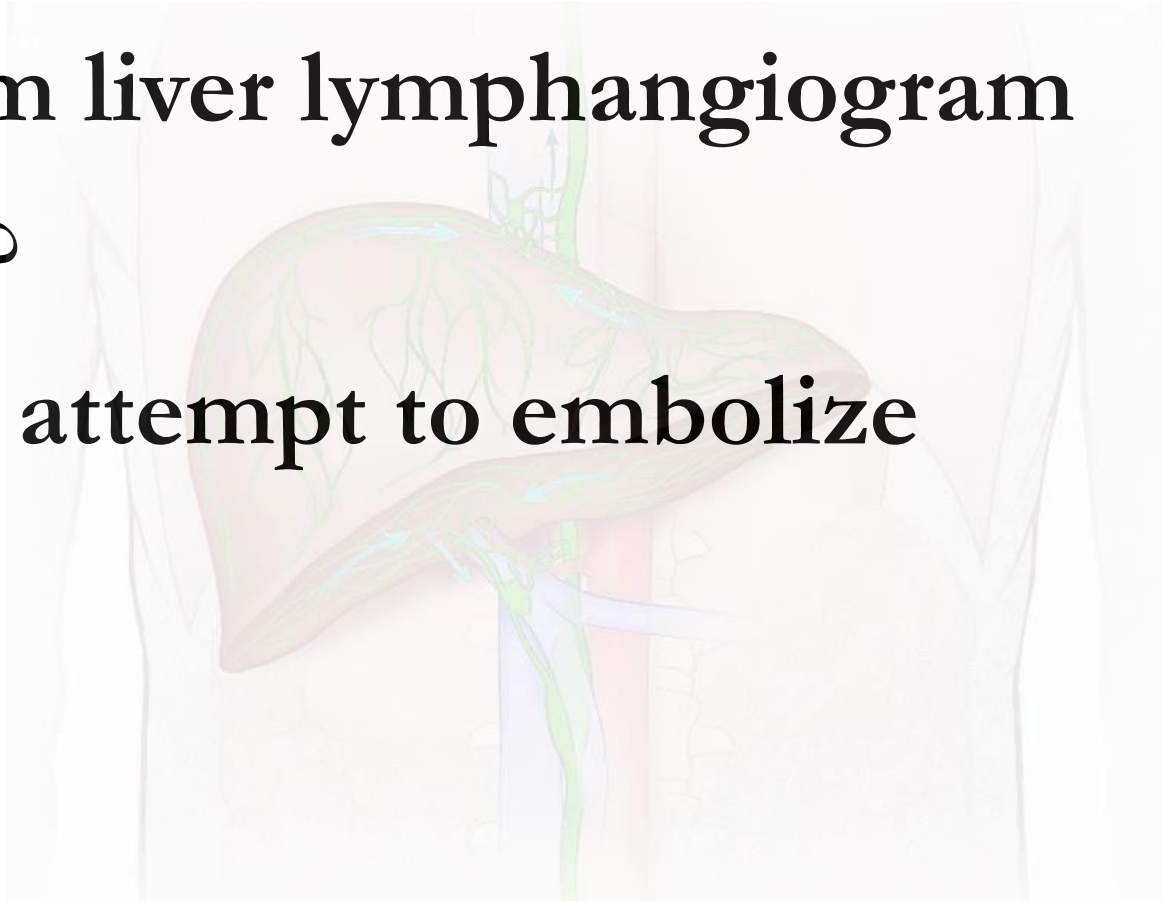
- The loss of the proteins in PLE happens from the liver lymph leaking into the intestine

PLE Treatment Concept

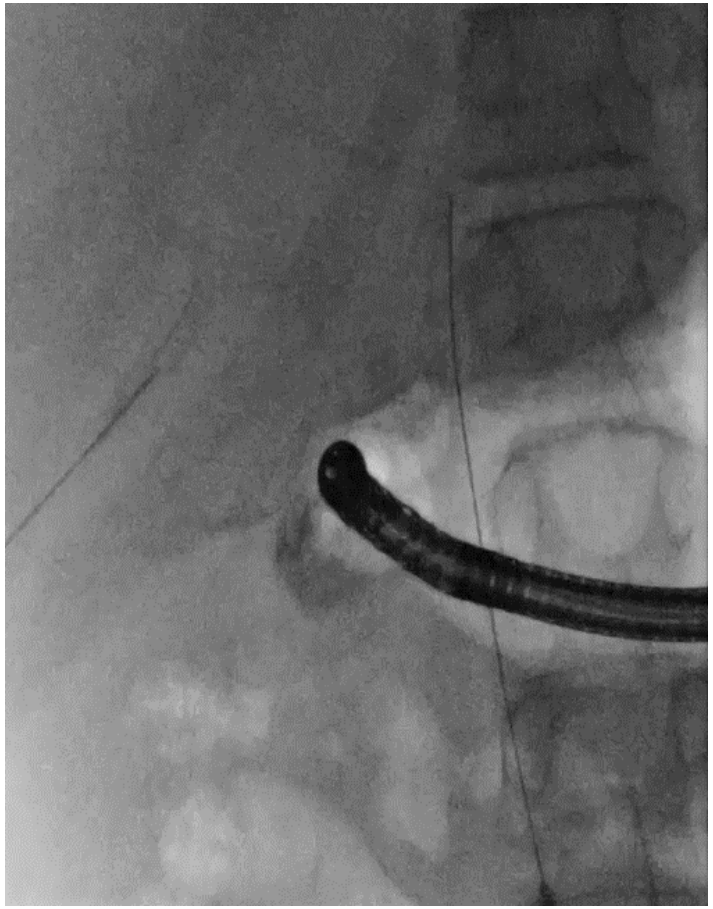
Perform liver lymphangiogram

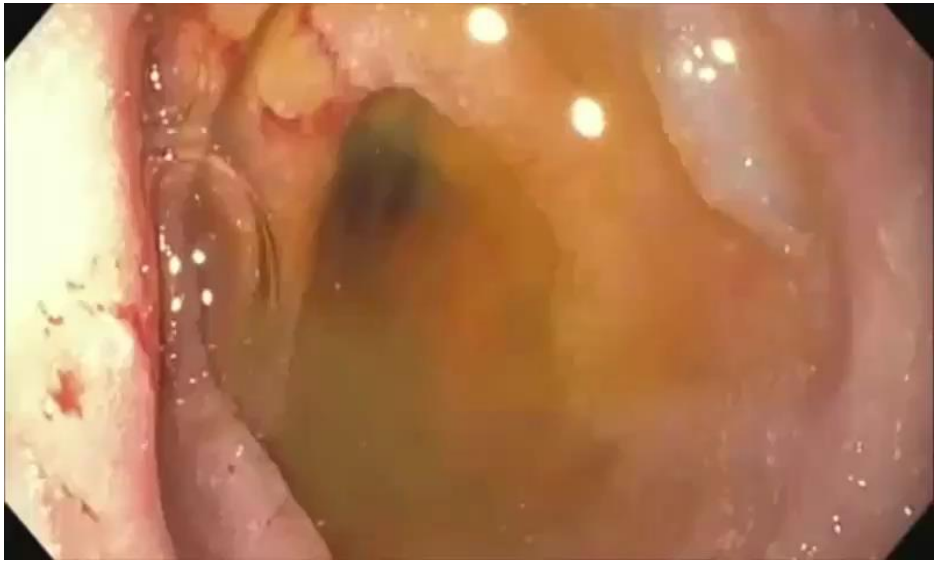
- Leak?

If leak attempt to embolize



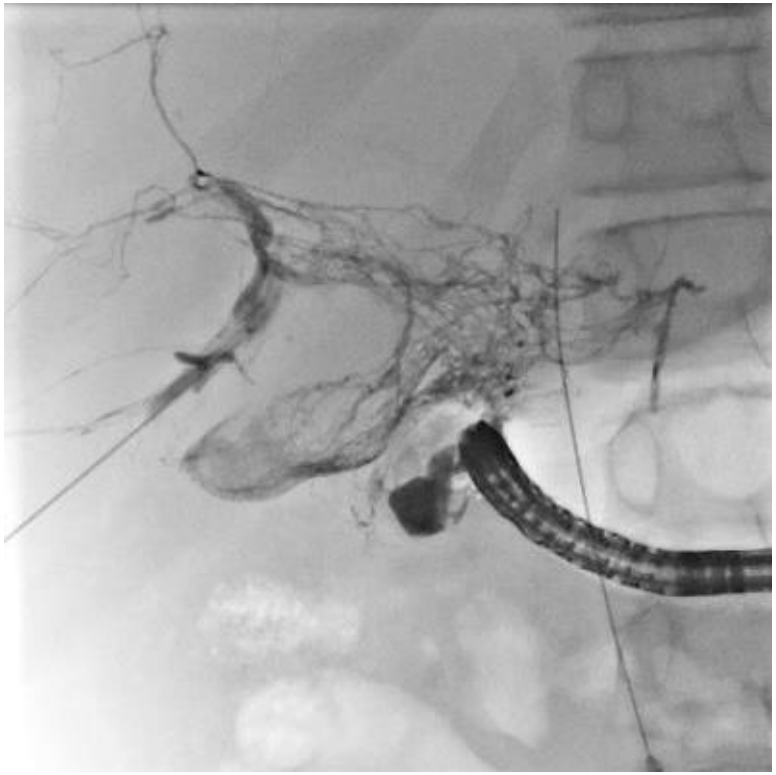
Liver Lymphangiogram PLE





Itkin M, Piccoli DA, Nadolski G, Rychik J, DeWitt A, Pinto E, et al. Protein-Losing Enteropathy in Patients With Congenital Heart Disease. J Am Coll Cardiol 2017;69:2929–37. doi:10.1016/j.jacc.2017.04.023.

Liver Lymphangiogram PLE



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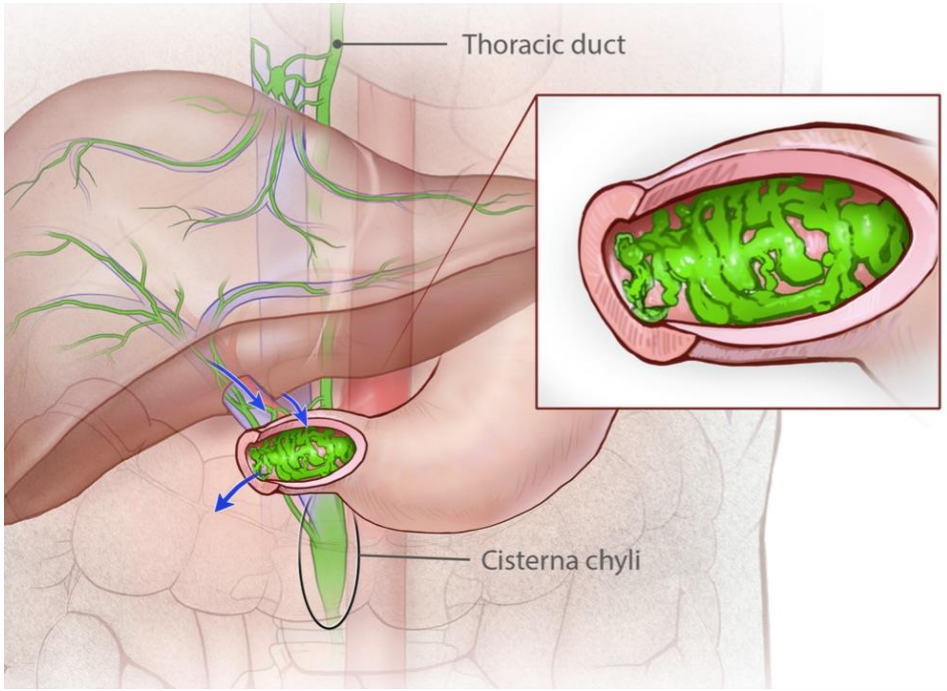
PLE-Leakage Diagram



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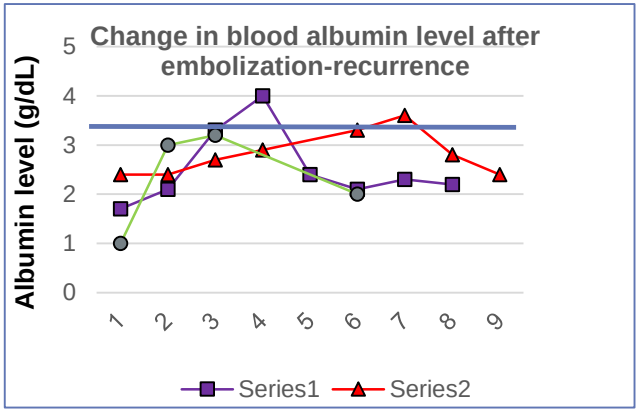
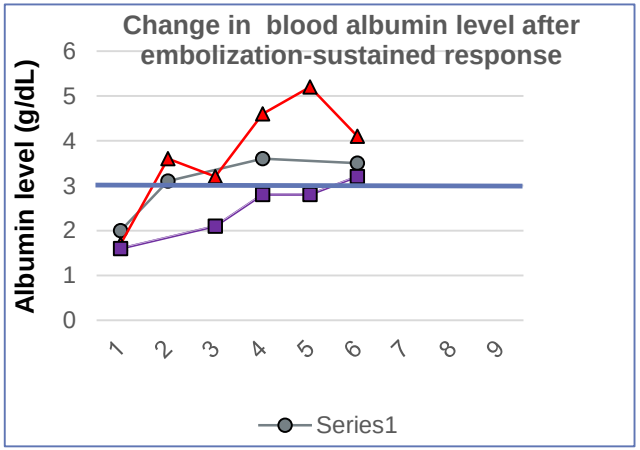
PLE Embolization Outcome

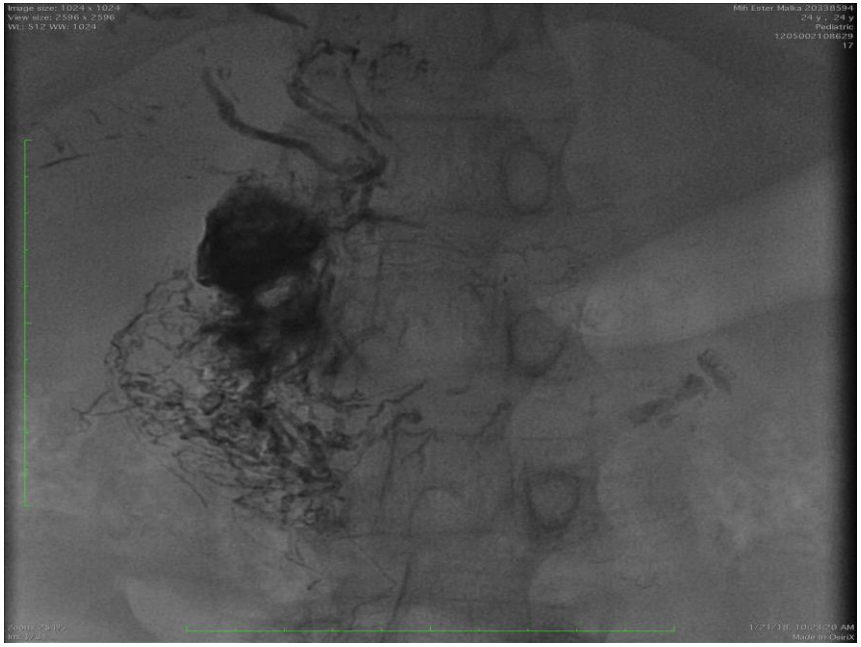
- 8 patients
- 3 patients temporary response
- 3 patients sustained response
- 363 days (range 84-1005)
- 2 duodenal bleeding

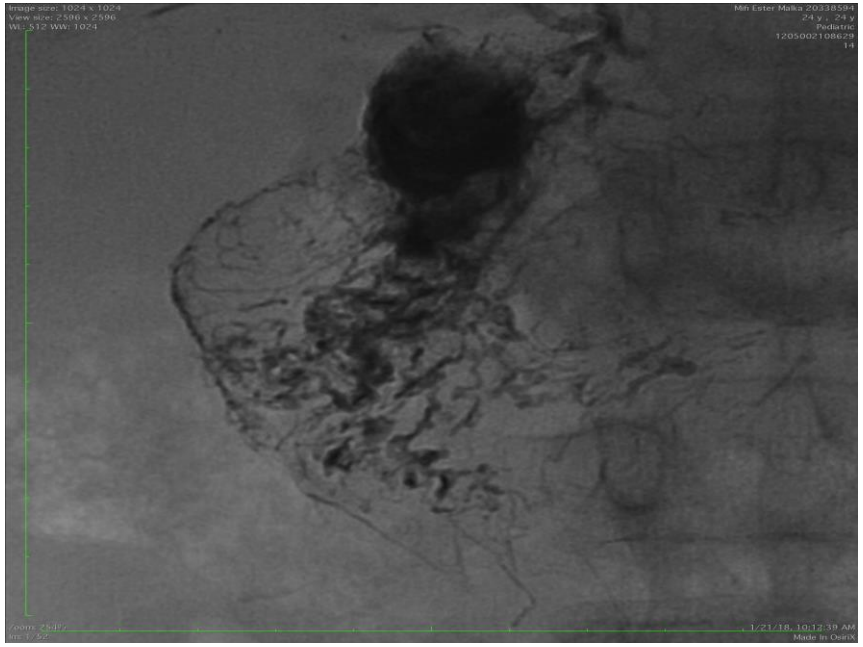




Itkin M, Piccoli DA, Nadolski G, Rychik J, DeWitt A, Pinto E, et al. Protein-Losing Enteropathy in Patients With Congenital Heart Disease. J Am Coll Cardiol 2017;69:2929–37. doi:10.1016/j.jacc.2017.04.023.

Embolization of Hepatoduodenal Communications





Conclusions

Lymphatic imaging is crucial in understanding of pathophysiology lymphatic

Understanding of the lymphatic variants is essential to explain some of the symptoms in patients with congenital heart disease

Improving the outcomes

Preventions and predictions


Thanks!
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